Journal of Surgical Oncology 64:341–346 (1997)

Primary Intrahepatic Biliary Cystadenomatous Tumors

D. TSIFTSIS, PhD, M. CHRISTODOULAKIS, MD, E. DE BREE, MD, AND E. SANIDAS, MD
Department of Surgical Oncology, University of Crete Medical School, Herakleion, Crete, Greece

Background: Biliary cystadenoma and cystadenocarcinoma are rare tumors. The clinical features of, and optimal surgical management for these lesions have not been defined clearly. In this report, we describe three cases of cystadenomatous tumors of the biliary tract: two of a cystadenoma and one of a cystadenocarcinoma. The differential diagnosis of the cystic tumors of the liver in countries with a high prevalence of liver hydatid disease is very important.
Methods: The authors report their institution's experience from 1988 to 1995 in treating two cystadenomas and one cystadenocarcinoma and review previously reported cases in the literature.
Results: Clinical presentation is usually mild and atypical. During operation, the mass was resected en bloc with a margin of normal liver tissue in the cases of cystadenomas and, in the case of cystadenocarcinoma, a left hepatectomy with drainage of the common bile duct was performed. The two patients with adenomas remain well at 1 and 7 years after operation and our patient with adenocarcinoma is free of disease 8 years later.
Conclusions: The treatment of choice is radical excision of the mass, either with a wide margin of normal liver or by means of a typical lobectomy, depending on the size and location of the lesion. When the lesion can be removed completely, the prognosis is excellent.
J. Surg. Oncol. 64:341–346, 1997.

KEY WORDS: **hepatic cystic tumors; biliary cystadenoma; biliary cystadenocarcinoma**

INTRODUCTION

Cystadenoma and cystadenocarcinoma are rare cystic tumors constituting fewer than 5% of all intrahepatic cysts of biliary origin [1]. They arise from the intrahepatic and rarely from the extrahepatic bile ducts and occur predominantly in middle-aged women. The most common presenting symptom is abdominal distension or an abdominal mass. The preoperative diagnosis is difficult. Surgical resection yields excellent results [2].

CASE REPORTS

Case 1

A 26-year-old woman presented with a 1-week history of epigastric pain, radiating to the back and aggravated by deep breathing. Laboratory tests showed slightly elevated bilirubin and ALT. Serological markers for hydatid disease were negative. Ultrasonography revealed a multilocular cystic mass on the lower surface of the left lobe of the liver (diameter 3.5 cm), with partial obstruction of the left hepatic duct, resulting in dilatation of the proximal biliary tree (diameter = 1.2 cm). Computed tomography (CT) demonstrated a cyst of the same diameter (Fig. 1) and dilation of the intrahepatic bile ducts of the left lobe. Findings were consistent with a hydatid cyst with possible communication to the biliary tree. On laparotomy, the cystic mass was found to be projecting through the visceral surface of the liver close to the ligament teres. The mass was resected *en bloc* with a margin of normal liver tissue. Cholecystectomy, exploration of the common bile duct and sphincteroplasty were performed. No stones were found, but the bile contained bile

*Correspondence to: Dr. Dimitris D. Tsiftsis, Department of Surgical Oncology, University General Hospital, P.O. Box 1352, Herakleion 711.10, Crete, Greece
Accepted 6 January 1997

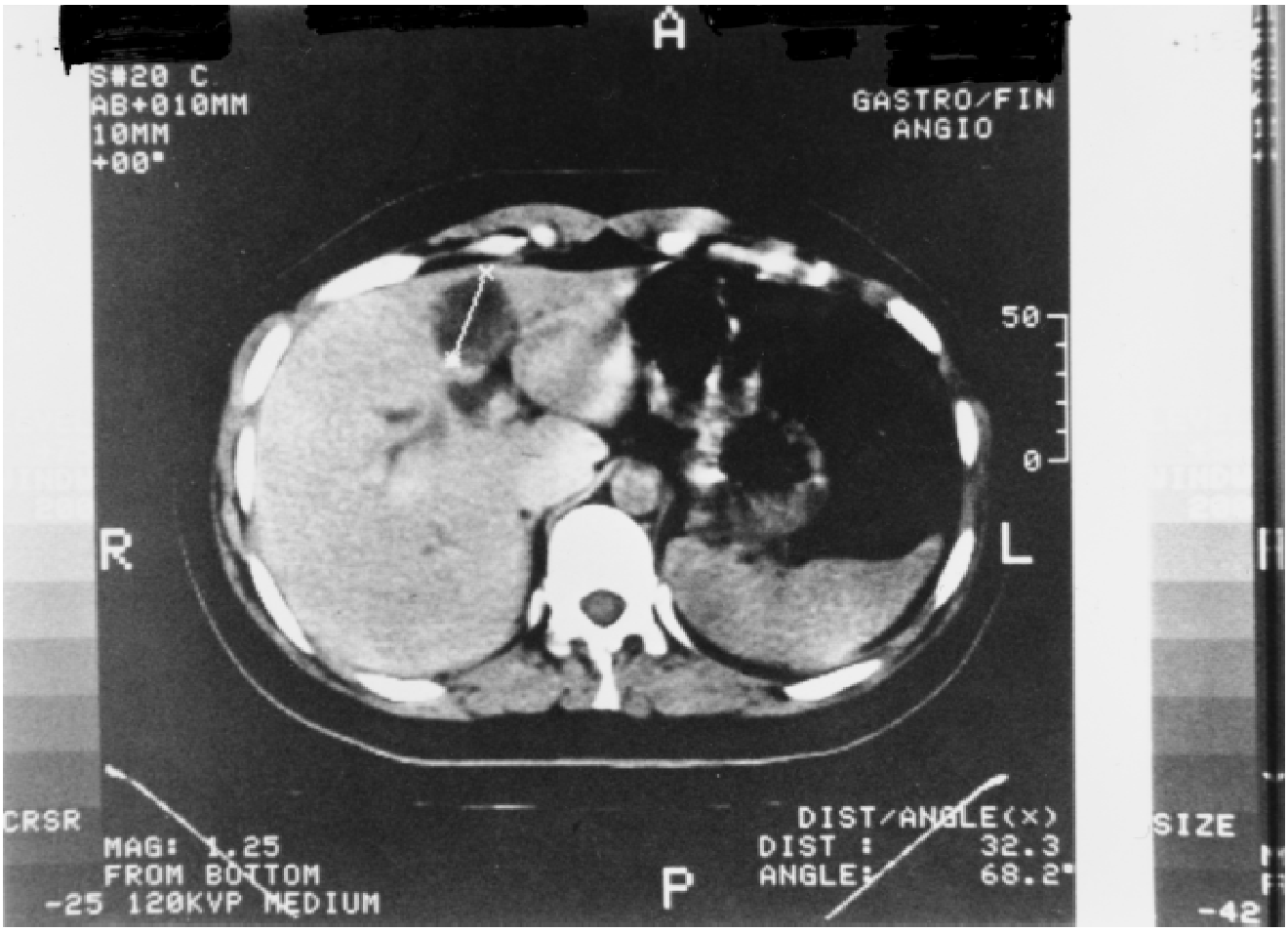

Fig. 1. CT scan. Biliary cystadenoma in the visceral surface of the left lobe of the liver causing dilation of the biliary ducts proximally.

macroaggregates. The postoperative course was uneventful. Histological examination demonstrated a biliary cystadenoma. She had no recurrence during a follow-up of 7 years.

Case 2

A 65-year-old woman was admitted to the medical ward with a painful epigastric mass. She reported a 6-kg weight loss during the previous 6 months, as well as mild epigastric pain and epigastric fullness. Laboratory tests were normal, and serological markers for hydatid disease were negative. Abdominal ultrasonography revealed a large multilocular mass 15 × 11-cm in the left lobe of the liver. CT of the abdomen confirmed the presence of a cystic mass with septa and solid components, giving the impression of a hydatid cyst (Fig. 2). During operation, the structure of the cystic wall and the content of the cyst raised the suspicion of malignancy and a left hepatectomy (Fig. 3) with drainage of the common bile duct was performed. Histological examination demonstrated a biliary cystadenocarcinoma (Fig. 4). The patient shows no signs of local or distant recurrence 8 years later.

Case 3

A 58-year-old woman presented with a 3-week history of right upper quadrant pain and nausea. Ultrasonography for suspected gallbladder disease revealed a septated cystic mass at the liver hilus. CT confirmed the presence of a well-encapsulated cystic mass (11 × 8 × 10 cm) between the right and left lobes of the liver, projecting inferiorly (Fig. 5). Laboratory tests were normal and serological markers for hydatid disease were negative. On laparotomy, a large encapsulated multiloculated cyst was found, involving both lobes of the liver, adherent to the gallbladder and duodenum. The mass was resected en bloc with the gallbladder and a margin of normal liver tissue. The common bile duct was not explored. Histological examination showed a biliary cystadenoma. Postoperatively, she developed a low-output external biliary fistula that healed in 30 days. She remains well 1 year after the operation.

DISCUSSION

Biliary cystadenomas are rare, slow-growing tumors with a propensity to become malignant. These tumors

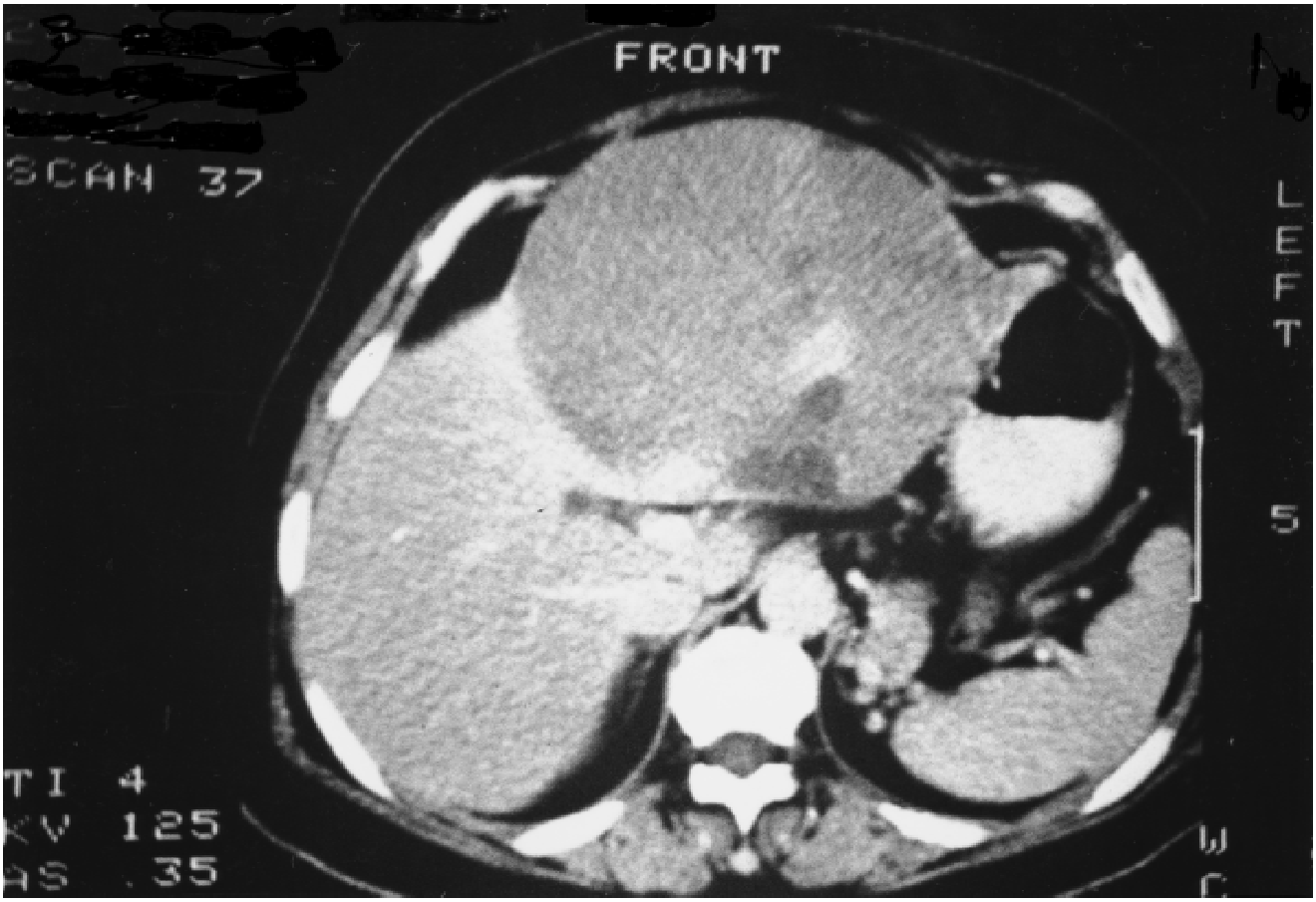

Fig. 2. CT scan. Biliary cystadenocarcinoma appearing as a big multilocular cyst in the left lobe.

comprise less than 5% of all intrahepatic cysts of biliary origin and 50% are in the right lobe, 29% in the left lobe, and 16% in both lobes. They are almost exclusively found in middle-aged women [3]. The size ranges from 1.5 to 35 cm, and they are lined with cuboidal epithelium and very often are multilocular with papillary infoldings. Biliary cystadenocarcinomas, the malignant counterpart, have a better prognosis than hepatocellular carcinoma or biliary cholangiocarcinoma [1]. Although they exhibit malignant characteristics such as mitotic activity, cell atypia, and infiltration, in places they retain the picture of a biliary cystadenoma (Fig. 6). The etiology of the tumors is largely unknown. One of the proposed theories is the development from ectopic remnants of primitive foregut sequestered within the liver [4].

Clinical presentation is usually mild and atypical. The duration of symptoms ranges from 3 days to 30 years. Many patients are asymptomatic, except for an increase in abdominal girth or the presence of a palpable mass. If symptoms occur, they include epigastric or right upper quadrant pain, nausea, jaundice, and a palpable mass. Late symptoms, due to recurrence or metastatic disease, include ascites, jaundice, and bone pain. The presenting symptom in one of our patients was jaundice, which is rare in the case of cystadenoma [5–7]. In the other cases, right upper quadrant pain and a palpable mass with weight loss were the presenting symptoms. Reported complications include hemorrhage, rupture, and inflammation.

The preoperative as well as the intraoperative diagnosis is difficult. The usual appearance on CT is of a single, multilocular low-density mass with papillary infoldings, and nodular thickening of the internal septa, that may show contrast enhancement [8]. Ultrasonography reveals an anechoic cystic mass with internal echoes representing papillary infoldings or a hypoechoic mass with echogenic septations. If calcification is present, the lesion can be mistaken for an echinococcal cyst [9]. The complementary roles of CT and ultrasonography in the diagnosis of cystic liver neoplasms has been extensively covered [10,11]. CT and angiography may give more information concerning the location and anatomic relation of the mass and may help in planning the operation. Ultrasonography and/or CT has been used for guidance in the percutaneous aspiration of the cystic fluid and needle biopsy [5]. A metastatic cystadenocarcinoma to the liver from a pancreatic or ovarian primary must be excluded with certainty. At operation, the cystic tumor is characterized by a thick wall and many septae lined by mucosa. Its content is viscous and yellowish in color.

The differential diagnosis includes congenital hepatic cysts, hydatid cyst, hepatic abscess, cystic hamartoma, hepatoma, primary or metastatic necrotic neoplasms, metastatic cystadenocarcinomas, Caroli's disease, post-

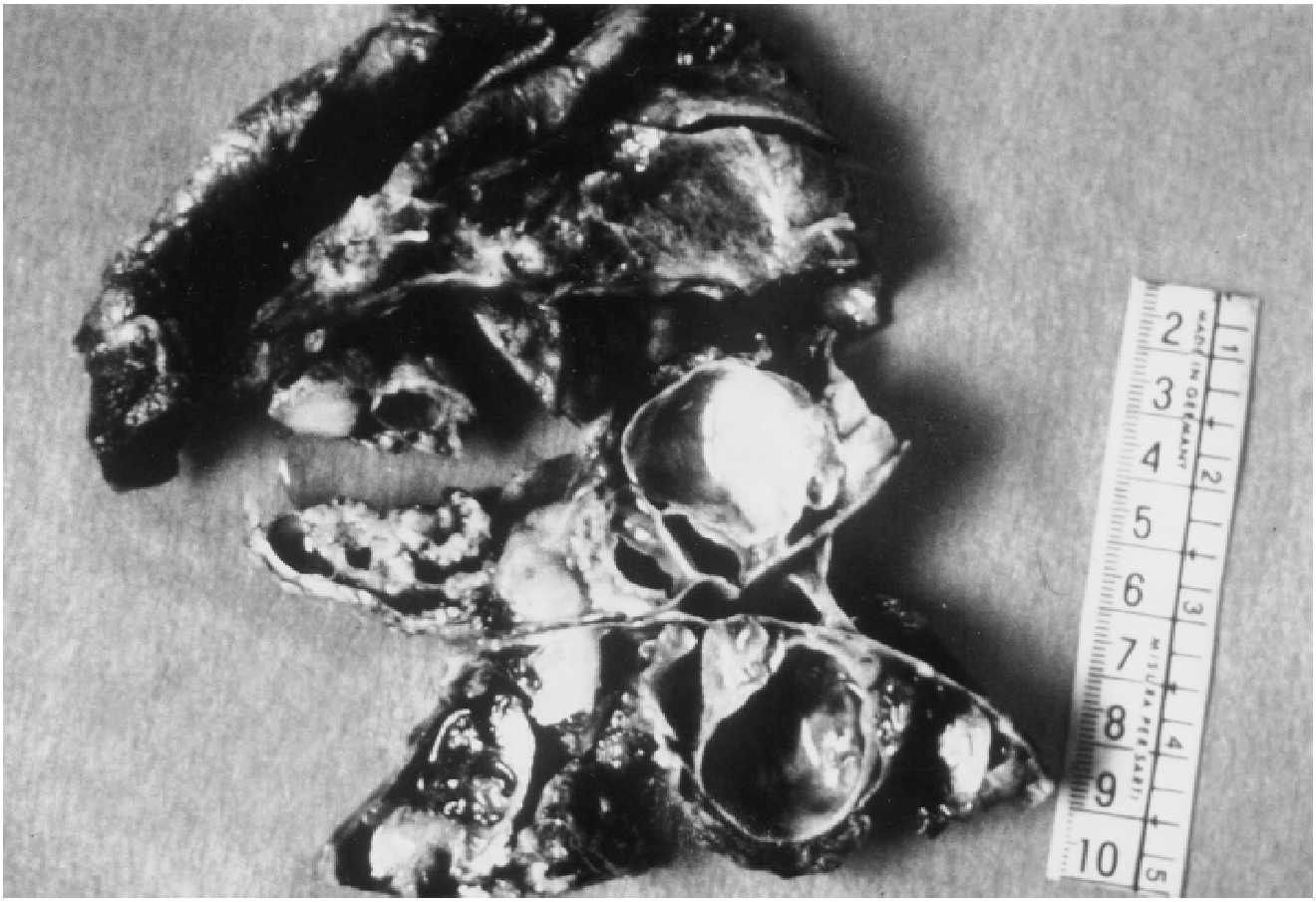

Fig. 3. The surgical specimen of a left lobectomy containing the biliary cystadenocarcinoma.

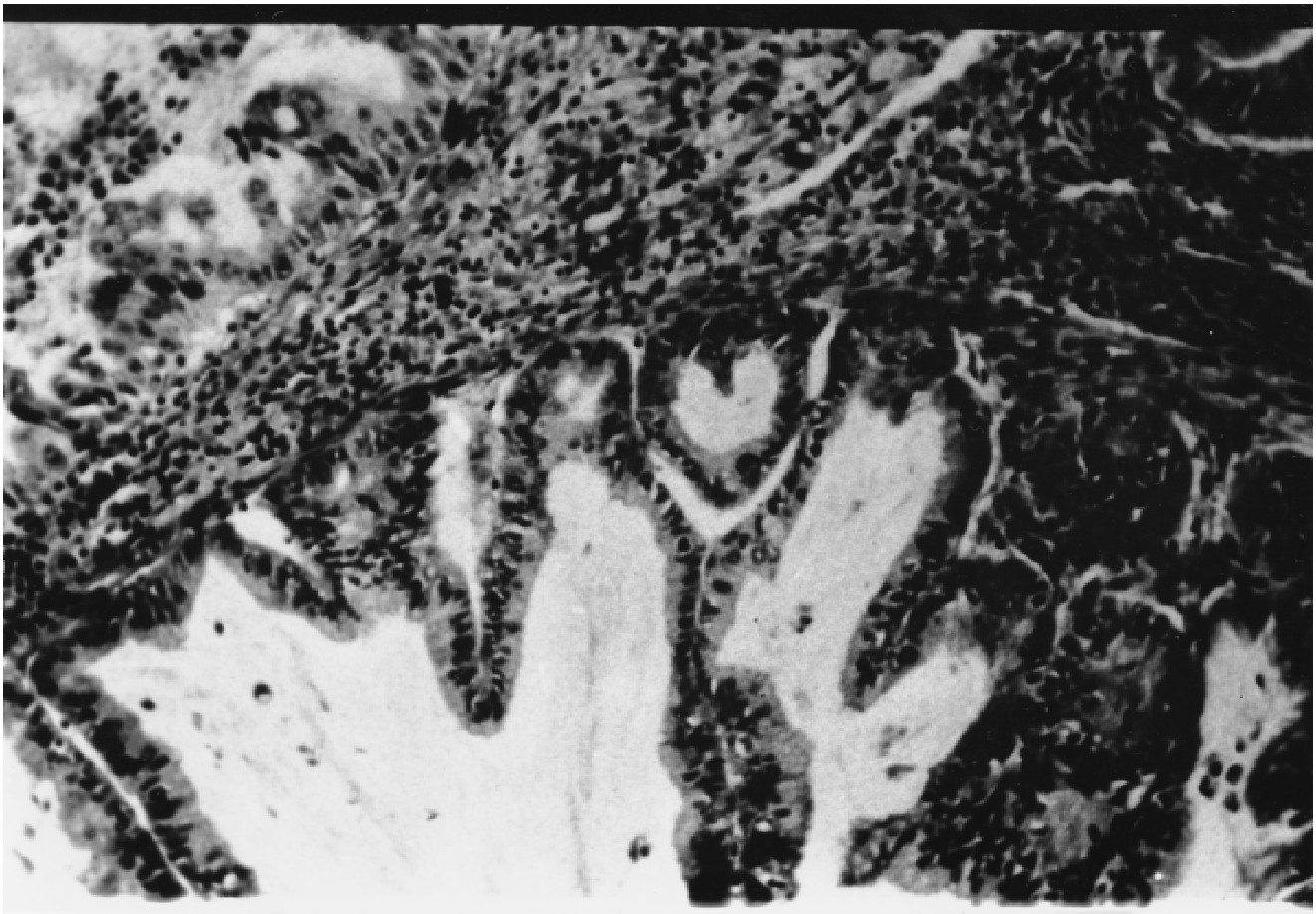

Fig. 4. Biliary cystadenocarcinoma. The lining of the cyst shows malignant characteristics

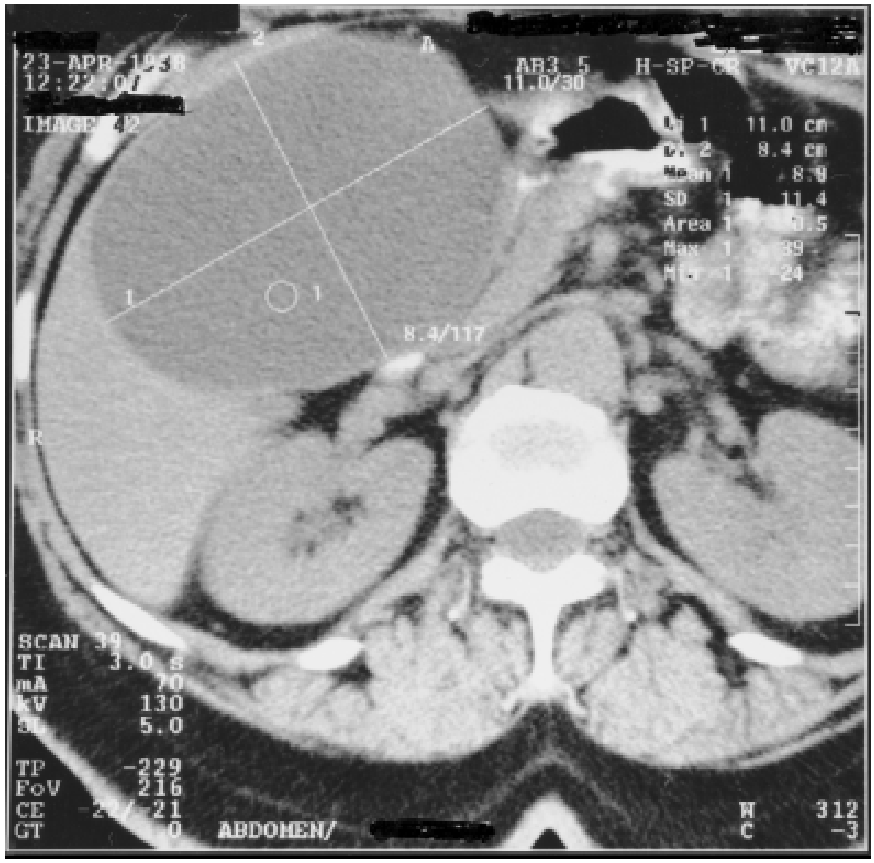

Fig. 5. CT scan shows a well-encapsulated cystic mass (11 × 8 × 10 cm) between the right and left lobes of the liver, projecting inferiorly.

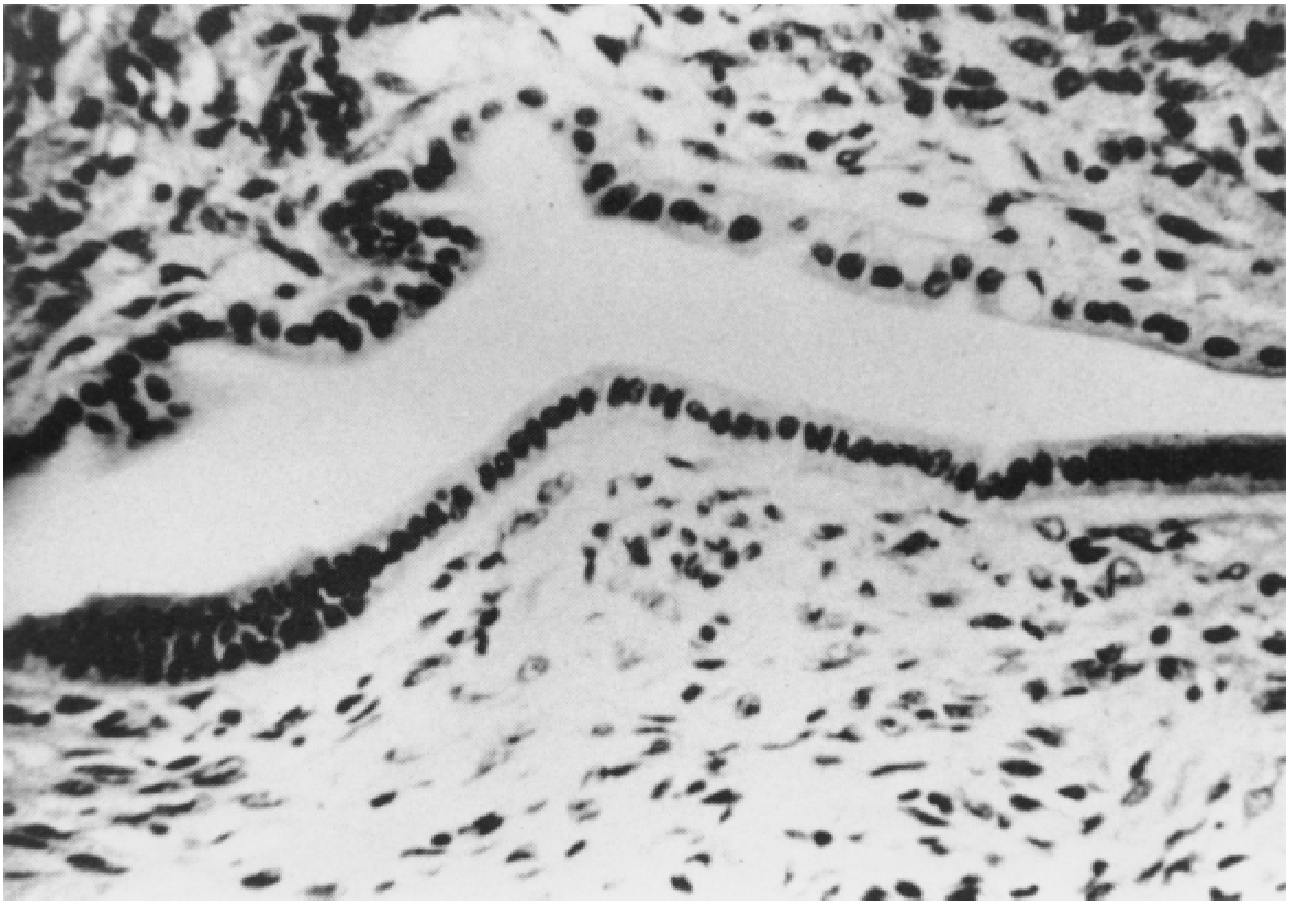

Fig. 6. Another site from the preview specimen with characteristics of benign biliary cystadenoma.

traumatic cyst, and polycystic disease [5,12,13]. In countries with a high prevalence of hydatid disease, the suspicion of a cystadenomatous tumor should always arise when negative serological tests are combined with a cyst that is septated whose content has a density higher than that of water on CT [14–16].

The treatment of choice is radical excision of the mass, either with a wide margin of normal liver or by means of a typical lobectomy, depending on the size and the location of the lesion [3,12,13]. Aspiration, sclerosis, marsupialization, and internal drainage must be avoided. Inadequate excision of both cystadenomas and cystadenocarcinomas will lead to recurrence in all cases [2]. Distal metastases are rare and are mainly to the liver, lungs, and bones [17].

The two patients with adenomas remain well 1 and 7 years after operation, and our patient with adenocarcinoma is free of disease 8 years later.

REFERENCES

1. Ishak KG, Willis GW, Cummins SD, Bullick AA: Biliary cystadenoma and cystadenocarcinoma: Report of 14 cases and review of the literature. Cancer 1977;38:322–338.
2. Lewis WD, Jenkins RL, Munson L, ReMine SG, Cady B, et al.: Surgical treatment of biliary cystadenoma—a report of 15 cases. Arch Surg 1988;123:563–568.
3. Takayasu K, Muramatsu Y, Moriyama N, et al.: Imaging diagnosis of bile duct cystadenocarcinoma. Cancer 1988;61:941–946.
4. Woods GL: Biliary cystadenocarcinoma. Case report of the hepatic malignancy originating in benign cystadenoma. Cancer 1981;47:2936–2940.
5. Case records of the Massachusetts General Hospital. Case 46. N Engl J Med 1985;313:125–1282.
6. Organ B, Petrak J: Biliary cystadenoma. South Med J 1984;77: 262–265.
7. Iemoto Y, Kondo Y, Fukamachi S: Biliary cystadenocarcinoma with peritoneal carcinomatosis. Cancer 1981;48:1664–1667.
8. Itai Y, Araki T, Furui S, Yashiro N, Ohtomo K, Io M: Computed tomography of primary intrahepatic biliary malignancy. Radiology 1983;147:485–490.
9. Papadimitriou J, Tsiftsis D, Tountas C: Hydatid cyst of the liver ruptured into the biliary tract. Curr Surg 1983;40:339–346.
10. Choi BI, Lim JE, Han MC, Lee DH, Kim YI, et al.: Biliary cystadenoma and cystadenocarcinoma: CT and sonographic findings. Radiology 1989;171:57–61.
11. Korobkin M, Stephens DH, Lee JKT, et al.: Biliary cystadenoma and cystadenocarcinoma: CT and sonographic findings. AJR 1989;153:507–511.
12. Federle M, Filly R, Moss AA: Cystic hepatic neoplasms: Complementary roles of CT and sonography. AJR 1981;136:456–348.
13. Frick MP, Feinberg SB: Biliary cystadenoma. AJR 1982;139:393–395.
14. Rutledge N, Pratt MC, Taupmann TE: Biliary cystadenoma mistaken for an echinococcal cyst. South Med J 1983;76:1575–1577.
15. Tsiftsis D, Sanidas E, Chatzakis G, Chroniaris N: Primary cystadenomatous tumors of the bile ducts. Eur J Surg Oncol 1992;18: 396–400.
16. Palacios E, Shannon M, Solomon C, Guzman M: Biliary cystadenoma: Ultrasound, CT, and MRI. Gastrointest Radiol 1990;15: 313–316.
17. Davies W, Chow M, Nagorney D: Extrahepatic biliary cystadenomas and cystadenocarcinoma. Ann Surg 1995;222:619–625.